Case Report

A Case of Behçet's Uveitis – A Delay in the Diagnosis

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Abstract

Behçet's uveitis is a severe ocular manifestation of Behçet's disease, a chronic, relapsing multisystem vasculitis. It typically presents as bilateral, recurrent, non-granulomatous panuveitis with occlusive retinal vasculitis involving both arteries and veins. Common ocular signs include hypopyon, hemorrhages, vitritis and retinal infiltrates. The underlying pathogenesis involves immune dysregulation in genetically predisposed individuals, often associated with HLA-B51.

Keywords: Behçet's, Vitritis, HLAB51 positive, Tofacitinib

Introduction

Behçet's disease is a chronic, relapsing, multi-systemic inflammatory disorder with a triad of symptoms which include oral ulcers, genital ulcers and uveitis. Uveitis in Behçet's disease is known to be bilateral, recurrent and non-granulomatous, which presents as panuveitis and retinal vasculitis in the eye. It is a potentially blinding condition and hence requires a high degree of clinical suspicion for early diagnosis, coupled with early and aggressive treatment to prevent blindness.[1]

Case Report

A 30-year-old male presented with complaints of diminution of vision in both eyes for 10 days, more pronounced in the left eye, associated with floaters and occasional redness. He reported a history of recurrent oral ulcers over the past 2 years. There was no history of trauma or prior similar ocular episodes. The patient had been previously diagnosed as a case of panuveitis elsewhere and had received oral corticosteroids and posterior sub-Tenon triamcinolone injection.

On examination, the best-corrected visual acuity (BCVA) was 6/60 in the right eye and PL + with inaccurate PR in the left eye. The patient was pseudophakic in both eyes. Pupillary examination revealed a sluggishly reacting pupil in the right eye and a non-reacting pupil in the left eye. Slit-lamp examination showed a quiet anterior chamber in both eyes, with no cells or flare. Intraocular pressure was within normal limits. Gonioscopy revealed open angles. The patient had developed oral ulcers, which pointed towards a presumptive diagnosis of Behçet's disease**.** There was no history of trauma or prior similar ocular episodes. Genital examination was normal.

Posterior segment examination revealed vitritis (4+), along with multiple intraretinal hemorrhages and perivascular sheathing suggestive of retinal vasculitis in both eyes, more severe in the left eye. The optic disc appeared pale.

Color fundus photography was done, which showed dense vitritis with marked media haze and obscuration of retinal details, consistent with severe posterior segment inflammation in the left eye and milder vitritis with visible optic disc pallor and perivascular sheathing of retinal vessels, suggestive of retinal vasculitis in the right eye.

Optical coherance tomography (OCT) was done, which revealed a distorted foveal contour with irregular retinal architecture and hyperreflective intraretinal areas, suggestive of inflammatory retinal involvement in the left eye.

Laboratory investigations revealed positivity for HLA-B51, while other infectious aetiology were ruled out.

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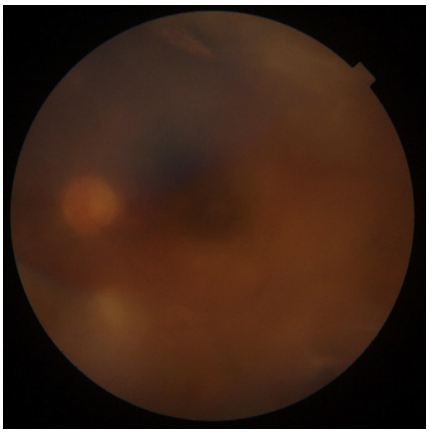

Figure 1: Vitritis of grade IV, presence of retinal hemorrhages along with disc pallor in the left eye

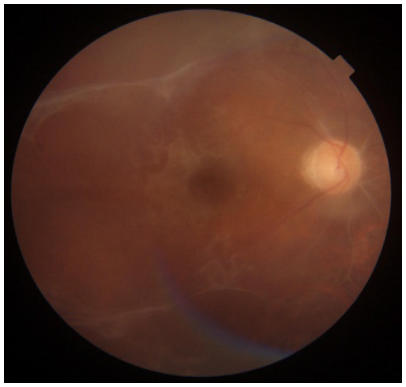

Figure 2: Mild vitritis of grade II with perivascular sheathing and traction bands in the right eye

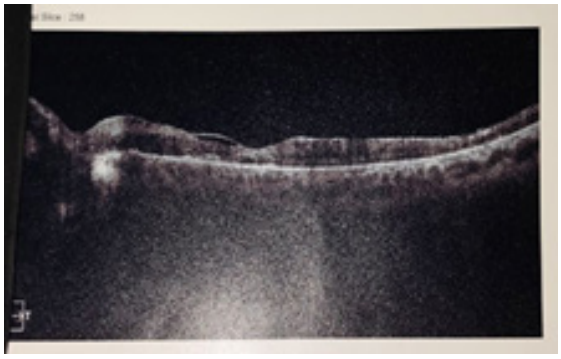

Figure 3: Distorted foveal contour with ERM and hyper reflective echoes in the vitreous

Based on the clinical findings and systemic features, a diagnosis of Behçet's uveitis was established. Following confirmation of the diagnosis, the patient was started on oral methylprednisolone (4 mg), tofacitinib (5 mg), and glutathione supplementation.

Discussion

This case highlights Behçet's disease presenting as posterior-predominant uveitis with a quiet anterior chamber, leading to initial misdiagnosis as panuveitis. The presence of recurrent oral ulcers and HLA-B51 positivity aided in establishing the diagnosis. Significant posterior segment involvement with vitritis, retinal vasculitis, and optic disc pallor underscores the aggressive nature of the disease (Figures 1-4).

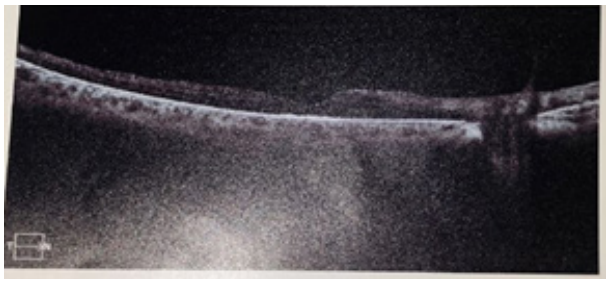

Figure 4: Normal foveal contour with ERM and few hyper reflective echoes in the vitreous cavity

Y. Wang *et al.* published a case of two young male patients presenting with panuveitis and recurrent oral and genital ulcers, who were initially misdiagnosed as Behçet's disease and treated with systemic corticosteroids with suboptimal response. Further evaluation revealed underlying syphilis and HIV infection. Following treatment with penicillin and antiretroviral therapy, uveitis resolved in one patient, while the other was lost to follow-up. This highlights the importance of screening for infectious etiologies in cases mimicking Behçet's disease.[2]

M Jari *et al***.** reported an 11-year-old girl who was initially diagnosed with preseptal cellulitis after presenting with pain and swelling around the left eye. A history of recurrent oral and genital ulcers was noted. On further evaluation, episcleritis and posterior uveitis were identified, and investigations revealed a positive pathergy test and HLA-B51, consistent with Behçet's disease. This case highlighted that ocular manifestations of Behçet's disease may be misdiagnosed due to overlapping clinical features.[3]

Conclusion

Behçet's uveitis is a potentially sight-threatening condition that may be misdiagnosed due to its variable clinical presentation. Ophthalmologists should be vigilant and maintain a high index of suspicion in patients presenting with uveitis, especially when associated with systemic features such as recurrent oral or genital ulcers. Early recognition of characteristic signs and timely initiation of appropriate therapy are essential to prevent irreversible visual loss.

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